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Primary Pericardial Synovial Sarcoma: A Case Report and Literature Review

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Primary synovial sarcoma of the heart is a rare tumor, with only six previous cases having been reported in the literature. Treatment has included surgery with or without chemotherapy. We present the first case of a documented synovial sarcoma arising from the pericardium in a 19-year-old man. Molecular analysis for t(X;18) SYT-SSX gene fusion was positive. Radiation treatment was given postoperatively to the entire heart with a boost to the area where the margins were positive.

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KEY WORDS: heart; pericardium; synovial sarcoma

INTRODUCTION

Primary cardiac tumors are extremely rare, ranging from 0.11% to 0.29% in various surgical series [1–3]. Approximately, 10%–17% of these are malignant [4–6], of which sarcoma is the most common. In order of frequency for cardiac sarcomas, these are angiosarcoma, fibrosarcoma, rhabdomyosarcoma, and malignant fibrous histiocytoma [7]. In general, synovial sarcoma has been rated as the fourth most common type of sarcoma affecting soft tissues, occurring in 5.6%–10% [8–11]. Only six cases to date have been reported, which primarily have arisen from cardiac tissue. None of these are known to be of pericardial origin. Our case represents the first known primary pericardial synovial sarcoma.

CASE REPORT

History

A 19-year-old east Indian male presented to the emergency department with a sudden onset of shortness of breath. A chest X-ray revealed a large heart, and a subsequent echocardiogram indicated a pericardial mass as well as a moderate pericardial effusion. Pericardiocentesis was performed with significant improvement in symptoms. An MRI revealed a 7 × 6 × 7.5 cm diffusely enhancing mass arising from the pericardium, extending anteriorly to the atrioventricular groove on the right side, with compression of the atrium; however, no direct cardiac invasion was noted. The lesion appeared to extend to the origin of the SVC superiorly and down to the diaphragm inferiorly, with no evidence of mediastinal or lung invasion (Figs. 1 and 2). Urgent thoracotomy was performed, which revealed a tumor mass arising from the inner surface of the pericardium. The tumor mass measured approximately 10 × 4 × 6 cm, with minimal direct extension to the right atrioinferior venocaval junction. Pericardial effusion cytology was negative for malignant cells. The tumor was excised and a partial pericardectomy was performed. Unfortunately, the margins were microscopically positive.

Pathology

The tumor was received in pieces attached to pericardium, all having a flattened, thick, sheet-like appearance with a gray-white color and areas of hemorrhage. The consistency varied from firm to soft and friable.

Microscopically, the tumor had a characteristic biphasic appearance of hypercellular spindled-cell sheets (Fig. 3) and well-differentiated glands (Fig. 4), some containing eosinophilic secretions. Both patterns demonstrated strong cytokeratin positivity as well as vimentin positivity in the spindled areas. Molecular analysis for t(X;18)

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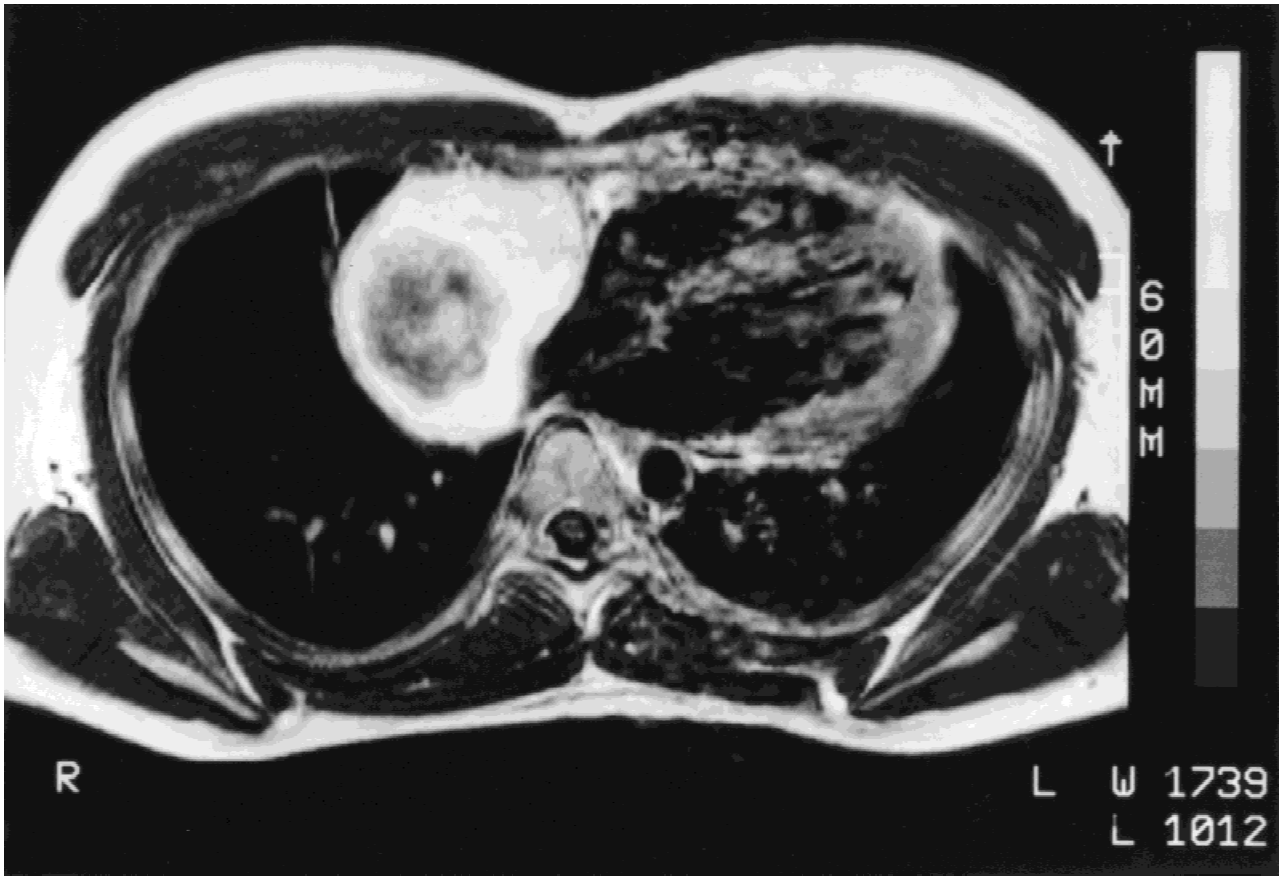

Fig. 1. Axial postenhancement T_1-weighted MRI showing an extensive mass relating to the right lateral pericardial surface (see also Fig. 2). The mass displaces the inferior part of the superior vena cava and decompresses the lateral aspect of the right atrium and runs into the inferior pericardial surface. Eccentrically within it an area of necrohaemorrhage is seen surrounded by a relatively uniformly enhancing mass of abnormal soft tissue.

Fig. 2. Coronal postenhancement T_1-weighted MR images showing an extensive mass relating to the right lateral pericardial surface. For detail, see legend for Figure 1.

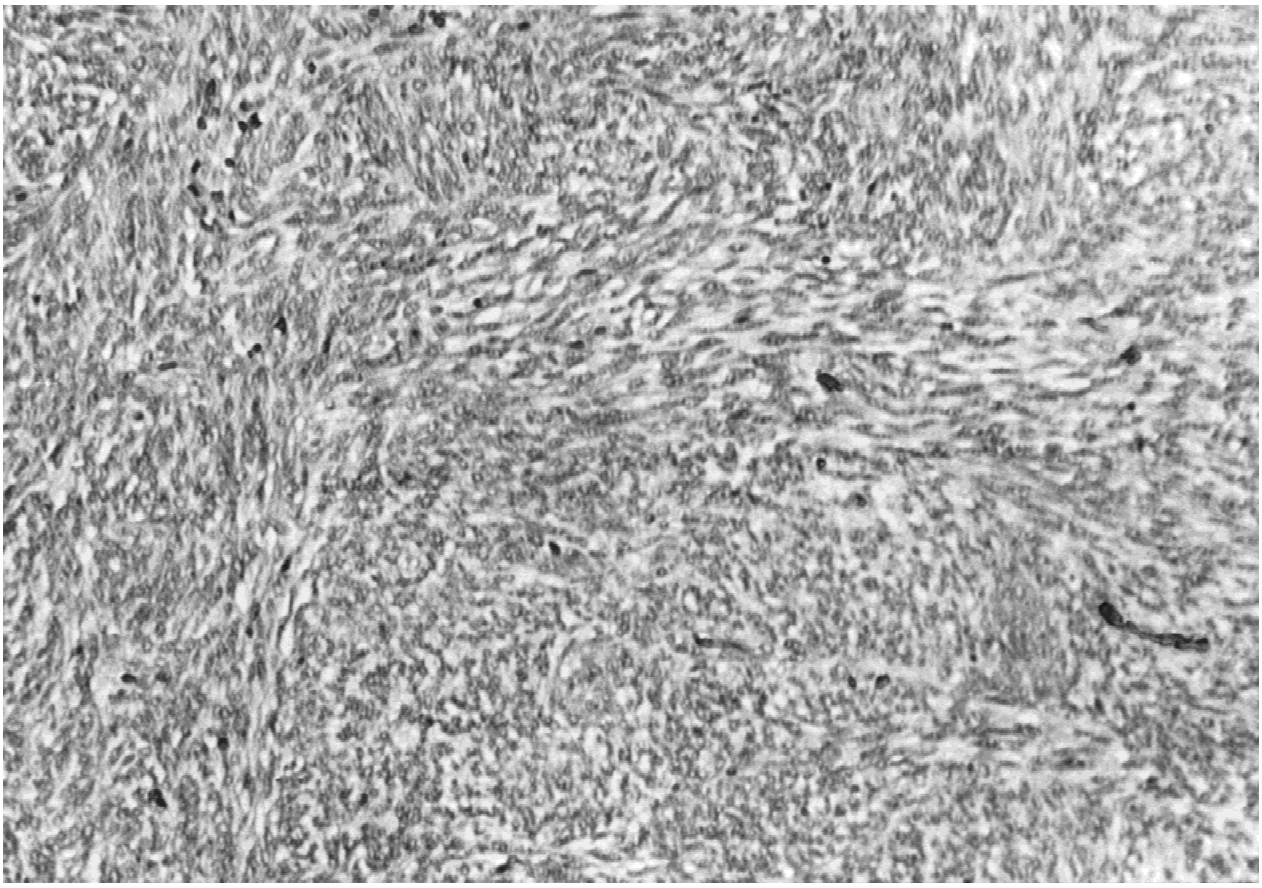

Fig. 3. Uniform hypercellular areas composed of spindle cells in the interglandular areas. (Hematoxylin-eosin stain.)

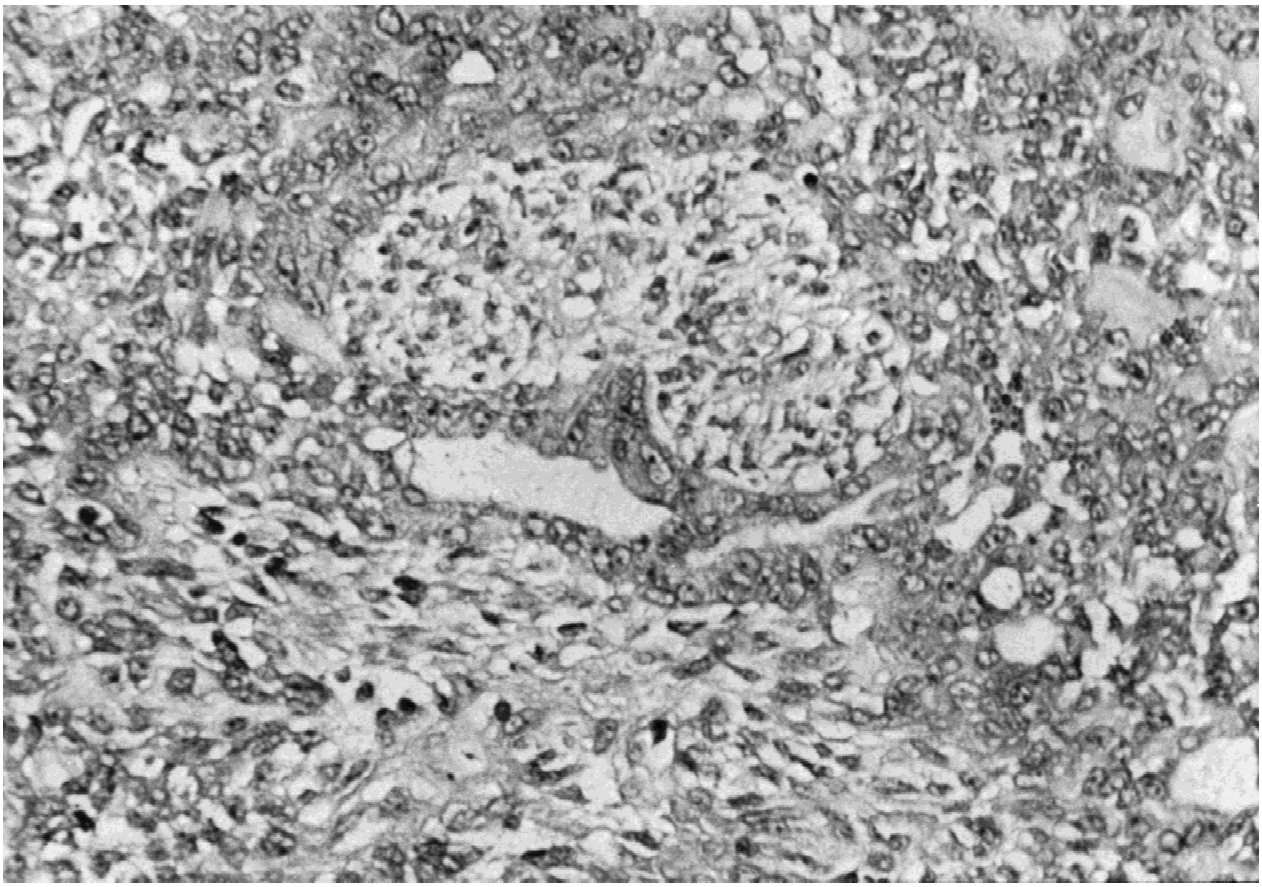

Fig. 4. An area of epithelial glandular differentiation. (Hematoxylin-eosin stain.)

TABLE I. Age, Sex, and Originating Sites of Primary Revicordial Synovial Sarcoma

Source	No. of Patients	Sex	Age	RA	RV	Interatrial septum	LA	LV	Pericardium
Burke et al. [12]	1	M	30		1				
	1	F	46				1		
Sheffield et al. [13]	1	M	53			1			
Siebenmann et al. [14]	1	F	31			1			
Karn et al. [15]	1	M	35		1				
Nicholson et al. [16]	1	M	13	1					
Al-Rajhi et al.	1	M	19						1
Total	7	5M/2F	Average 32	1	2	2	1	0	1

TABLE II. Treatment and Survival

Source	Primary treatment	Adjuvant treatment	Status
Burke et al. [12]	None	None	Autopsy diagnosis
	Unknown	Unknown	Unknown
Sheffield et al. [13]	Surgery	None	Died at 6 months
Siebenmann et al. [14]	Heart transplantation	None	Died at 3 months
Karn et al. [15]	Surgery	Doxorubicin, ifosfamide, and mesna	Died at 9 months
Nicholson et al. [16]	Surgery	Ifosfamide, vincristine, and actinomycin	Alive at 10 months
Al-Rajhi et al.	Surgery	RT to whole heart plus boost to involved site	Alive at 12 months with no evidence of disease

SYT-SSX gene fusion were positive, confirming the diagnosis of biphasic synovial sarcoma (molecular analysis was performed courtesy of Dr. M. Ladanyi, Memorial Sloan-Kettering Cancer Center).

Treatment

The patient was treated with a course of postoperative radiation therapy, given in two phases. When initially simulated in the supine position, the amount of lung within the treatment field was felt to be excessive, hence the patient was treated in a sitting position resulting in a reduced area of radiated lung. The patient was immobilized in this sitting position using a custom-made device with a vacuum bag posteriorly and a perspex shell anteriorly. The whole cardiac silhouette was treated with an anterior and posterior, parallel opposed field, using 6-MV photons. A dose of 3,060 cGy in 17 fractions was prescribed to the midplane over 3.5 weeks. Following this, using the shrinking field concept, an additional 1,980 cGy was delivered in 11 fractions over 2.5 weeks using a CT-based treatment plan to boost the tumor bed. For this part of the treatment, the patient was in the supine position.

DISCUSSION

Primary synovial sarcoma of the heart and pericardium is a rare event, arising mainly from the right side of the heart. Demographic characteristics and distributions are represented in Table I. Primary treatment has consisted of a surgical excision in most cases. Due to the rarity of this malignancy, no standardized adjuvant therapy is currently followed. Previously reported treatment modalities are summarized in Table II. Two previous cardiac cases have received adjuvant ifosfamide-based combination chemotherapy with no long-term survival reported. In the present case, the tumor appeared to be localized to the pericardium, and surgical excision was performed; however, the margins were microscopically involved.

Radiation therapy is known to be of value in cases where the margins are positive, to reduce local recurrence rates. Cardiac irradiation can lead to long-term cardiac damage as shown in the treatment of Hodgkin's disease [17,18]. The threshold tolerance of a 5% chance of late sequelae at 5 years (Td 5/5) for the whole heart is 4,500 cGy and for less than 1/3 rd of the heart is 5,000 cGy in conventional fractionation. In this case, postoperative radiation therapy was advised due to the microscopically involved margins. As there was a strong family history of cardiovascular problems, we restricted the total dose of radiation for fear of long-term toxicity.

The natural history of primary cardiac synovial sarcoma indicates a very poor survival, with most patients dying within a 6-month time period. However, for tumor of pericardial origin, the survival remains unknown. In this case, after surgical excision and postoperative adjuvant radiotherapy, the patient remains clinically free of disease at 12 months.

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